

An exceptional man

Stephen Hawking has survived almost 40 years with a disease that usually kills people 14 months after diagnosis. **Roger Dobson** asks why

Stephen Hawking developed motor neurone disease when he was in his early 20s. Most patients with the condition die within five years, and according to the Motor Neurone Disease Association, average life expectancy after diagnosis is 14 months.

But Professor Hawking, the Cambridge University physicist and cosmologist and author of *A Brief History of Time*, has confounded the statistics and recently celebrated his 60th birthday. No one is thought to have survived for so long with the incurable condition, which kills three people a day in the United Kingdom.

"Stephen Hawking is a fascinating case, and neurologists always puzzle over it. The case is fascinating because of the early onset and the length of time the disease has run," one neurologist said.

Motor neurone disease (MND), or amyotrophic lateral sclerosis, is a progressive, usually fatal neuromuscular disease. It attacks motor neurones in the spinal cord and lower brain, which transmit signals from the brain to the voluntary muscles throughout the body.

"The average duration of survival from diagnosis is about 14 months, but it varies enormously," says Professor Nigel Leigh, professor of clinical neurology at King's College, London, and director of the King's MND Care and Research Centre.

"We have found that the survival in younger patients is strikingly better and is measured in many years—in some cases more than 10. Among people in their 50s and 60s, there is a 50% chance of surviving four years or so. It is a different beast if you start young, oddly, and no one knows why. But even some forms of MND that start in the [patient's] 50s or 60s can be slowly progressive," he added.

"I have no personal knowledge of Stephen Hawking, but he is exceptional. I am not aware of anyone else who has survived with MND as long. What is unusual is not only the length of time, but that the disease seems

to have almost burnt out. He appears to be relatively stable, and I have had one or two patients where there is still a gradual deterioration, but where the curve has flattened off. In these cases MND started quite early, in the [patient's] 20s or 30s. This kind of stabilisation is extremely rare.



Stephen Hawking believes motor neurone disease is a syndrome that can have different causes. "Maybe my variety is due to bad absorption of vitamins"

"In early onset patients there appear to be biological differences. It is already clear that if you look at the genetics of MND, there are at least half a dozen, maybe a dozen, genetic forms of true MND. Another possibility is that there is some kind of interaction with the ageing process."

Professor Pam Shaw, professor of neurology at the University of Sheffield, said: "The older you are the quicker the disease course tends to be, but we don't really have a handle on why some people survive for longer periods than others. I wish we did."

Asked by the *BMJ* if he knew why his condition had evolved differently from a typical case of MND, Professor Hawking replied, "I believe motor neurone

disease is a syndrome that can have different causes. Maybe my variety is due to bad absorption of vitamins."

Professor Hawking supplements his diet with daily mineral and vitamin tablets, and zinc, cod liver oil capsules, folic acid, vitamin B complex, vitamin B-12, vitamin C and vitamin E are said to have been particularly helpful. He also follows a diet free of gluten and vegetable oil and avoids convenience foods; quite recently he started to include a small amount of dairy produce.

As far as medical care is concerned, he receives passive chest physiotherapy and passive and

"I was in for two weeks, during which I had a wide variety of tests. After all that, they didn't tell me what I had, except that it was not multiple sclerosis, and that I was an atypical case. I gathered, however, that they expected it to continue to get worse, and that there was nothing they could do, except give me vitamins. I could see that they didn't expect them to have much effect. I didn't feel like asking for more details, because they were obviously bad.

"The realisation that I had an incurable disease that was likely to kill me in a few years was a bit of a shock."

He later married Jane Wilde and they had three children. His condition gradually deteriorated but he managed to cope with the help of his wife and research students until 1980 when he changed to a system of community and private nurses.

"This lasted until I caught pneumonia in 1985. I had to have a tracheostomy operation. After this, I had to have 24 hour nursing care, made possible by grants from several foundations... The tracheostomy operation removed my ability to speak altogether.

"For a time, the only way I could communicate was to spell out words letter by letter, by raising my eyebrows when someone pointed to the right letter on a spelling card. It is pretty difficult to carry on a conversation like that, let alone write a scientific paper."

"A computer expert in California, Walt Wolosz, heard of my plight. He sent me a computer program he had written. This allowed me to select words from a series of menus on the screen, by pressing a switch in my hand. The program could also be controlled by a switch, operated by head or eye movement. When I have built up what I want to say, I can send it to a speech synthesiser.

"I have had motor neurone disease for practically all my adult life. Yet it has not prevented me from having a very attractive family, and being successful in my work. This is thanks to the help I have received from Jane, my children, and a large number of other people and organisations. I have been lucky, that my condition has progressed more slowly than is often the case. But it shows that one need not lose hope." □